

Letter to Editor regarding “Waxing and Waning Presentation of Isolated Cardiac Sarcoidosis on Sequential 18F-FDG-PET Exams”

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I read with interest the article Ostwani et al (1) and am concerned that the patient does not have cardiac sarcoidosis (CS) for three reasons. Firstly, because of the lack of a positive biopsy and therefore the patient does not meet guideline criteria for the diagnosis of CS.(2) Secondly, because corticosteroid refractory sarcoidosis is considered very rare.(3) The third reason for concern is that the accumulating data suggests that there may not be a pathophysiological entity of truly isolated CS. It is clear that there are many patients, with manifest CS, who have no clinically apparent disease in other organs i.e. can be termed *clinically* isolated CS. However, sarcoidosis is, by definition and biology a systemic disease. Hence a key starting point to understand isolated CS is to agree on a standardized definition. The 2017 version of the Japanese CS guidelines tackled, for the first time, the definition of and criteria for the diagnosis for isolated CS.(4) They included the following three criteria:

- i) No clinical findings characteristic of sarcoidosis are observed in any organs other than the heart.
- (ii) ⁶⁷Ga scintigraphy or ¹⁸F-FDG PET reveals no abnormal tracer accumulation in any organs other than the heart.
- (iii) A chest CT scan reveals no shadowing along the lymphatic tracts in the lungs or no hilar and mediastinal lymphadenopathy (minor axis >10 mm).

Using a similar definition, we found *imaging* isolated CS in only one in 31 cases.(5) However, other data suggest that even these apparent isolated cases are unlikely to be truly isolated. Petek et al investigated 10 patients with presentations and cardiac imaging consistent with the Japanese definition of isolated CS. Four of these 10 had granulomas on bronchial biopsy.(6) Hence these data suggest that there is a small subset of patients who at the moment of FDG-PET imaging have 'PET detectable inflammation' *only* in their heart. However, it also follows, that additional or interval investigation will likely reveal extra-cardiac disease.

This debate is more than just semantics as the overdiagnosis of 'imaging isolated CS' can, as in this case(1), lead to unnecessary immunosuppression and/or 'missed' alternative diagnosis.

References

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